

# Clinic experience in discoid lupus erythematosus: a retrospective study of 132 cases

Goknur Ozaydin Yavuz<sup>1</sup>, Ibrahim Halil Yavuz<sup>1</sup>, Irfan Bayram<sup>2</sup>, Rojda Aktar<sup>1</sup>, Serap Gunes Bilgili<sup>1</sup>

<sup>1</sup>Department of Dermatology, Faculty of Medicine, Van Yuzuncu Yil University, Van, Turkey

<sup>2</sup>Department of Pathology, Faculty of Medicine, Van Yuzuncu Yil University, Van, Turkey

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## Abstract

**Introduction:** Lupus erythematosus (LE) is an autoimmune disease characterized by a broad range of cutaneous manifestations. Discoid LE (DLE) is the most common chronic manifestation of LE. Literature reviews show that there are a limited number of large-series studies investigating DLE. Additionally, there is still no consensus on the etiological factors of DLE such as sun exposure and smoking.

**Aim:** To evaluate the clinical and demographic characteristics of patients with DLE.

**Material and methods:** The study included patients who were hospitalized in the inpatient and outpatient clinics at the Dermatology Department. Age, gender, treatment method, history of smoking, antinuclear antibody positivity, progression to systemic lupus erythematosus (SLE), photosensitivity, and laboratory findings were recorded for each patient.

**Results:** The study included 132 patients comprising 67 (50.8%) men and 65 (49.2%) women. A family history was found in 3.8%, SLE was detected in 5.3%, and photosensitivity was revealed in 50.0% of the patients. ANA positivity was found in 23.7%, a history of smoking was revealed in 61.4%, and chronic sun exposure was detected in 42.4% of the patients.

**Conclusions:** Discoid LE, though identified long ago, remains unelucidated and there are very few studies in the literature reporting on DLE. The results indicated that smoking and chronic UV exposure are important risk factors for DLE. Moreover, although ANA positivity was high in our patients, the rate of progression to SLE was remarkably low. The results also showed that, contrary to common belief, there is no female preponderance in DLE.

**Key words:** discoid lupus erythematosus, smoking, antinuclear antibody.

## Introduction

Lupus erythematosus (LE) is an autoimmune disease characterized by a broad range of cutaneous manifestations. After rheumatologic manifestation, cutaneous lupus erythematosus (CLE) is the second most common clinical manifestation of LE [1–4]. There are several distinct clinical presentations of CLE, mainly including acute CLE, subacute CLE (SCLE), and chronic CLE. Moreover, discoid LE (DLE) is another form which is the most common manifestation of CLE [2].

Discoid LE (DLE) is mostly seen in women in their 5<sup>th</sup> and 6<sup>th</sup> decades of life. DLE usually has a more benign course as compared to other CLE subtypes. Discoid LE often involves the head and neck, predominantly the hairy areas of the skin such as scalp. Involvement of hairy skin is seen in 60% of patients with DLE while isolated involvement of hairy skin is seen in only 10% of patients.

A previous study also reported that scarring alopecia was detected in 34% of patients with DLE [1, 3].

The etiopathogenesis of DLE is multifactorial. Of note, genetic and environmental causes and congenital and acquired immune response change have been reported as the leading causes of DLE. Moreover, ultraviolet (UV) exposure is regarded as the most common cause of DLE, followed by drugs, smoking, vitamin D deficiency, and HIV infection [1, 4–6]. On the other hand, 5–10% of patients with DLE may progress to systemic lupus erythematosus (SLE), which shows that the localized form of DLE is not the only form [1].

The primary step in the treatment of DLE is prevention of the disease, which is dependent on a combination of patient education and avoidance of predisposing factors. Mainstay treatment includes topical steroids, topical calcineurin inhibitors, and antimalarial drugs [6, 7].

**Address for correspondence:** Goknur Ozaydin Yavuz MD, Department of Dermatology, Faculty of Medicine, Van Yuzuncu Yil University, 65100 Van, Turkey, phone: +90 5054753362, +90 5054753362, fax: +90 5054753362, e-mail: [goknuroz1@gmail.com](mailto:goknuroz1@gmail.com)

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Although DLE was identified long ago, there are a limited number of large-series studies in the literature. Additionally, there is still no consensus on the etiological factors such as sun exposure and smoking.

## Aim

In this study, we aimed to evaluate the clinical and demographic characteristics of patients with DLE.

## Material and methods

The retrospective study included patients who were hospitalized in the inpatient and outpatient clinics at the

**Table 1.** Demographic characteristics of patients

Parameter	Value	Percentage
Gender:		
Male	67	50.8
Female	65	49.2
Occupation:		
Farmer	20	15.2
Housewife/unemployed	72	54.5
Civil servant or other	40	30.3
Age [years]*	43.2 ±14.1	42.5
Age at onset [years]*	39.3 ±14.1	38.0

\*For quantitative values, mean ±SD was used instead of n and median was used instead of %.

**Table 2.** Clinical features of patients

Parameter		Number	Percentage
Family history	No	127	96.2
	Yes	5	3.8
SLE	No	125	94.7
	Yes	7	5.3
Photosensitivity	No	66	50.0
	Yes	66	50.0
ANA positivity	No	100	76.3
	Yes	31	23.7
Smoking	No	51	38.6
	Yes	81	61.4
Chronic sun exposure	No	76	57.6
	Yes	56	42.4
Comorbidity	DM	9	6.8
	Hyperthyroidism	3	2.3
	HT	24	18.2
	No	96	72.7

SLE – systemic lupus erythematosus, DM – diabetes mellitus, HT – hypertension, ANA – antinuclear antibody.

Medical School Dermatology Department due to DLE between January 2005 and December 2017. Medical records of the patients were obtained from hospital databases and then analyzed for age, gender, lesion site, treatment method, comorbidities, family history, history of smoking, antinuclear antibody (ANA) positivity, progression to SLE, photosensitivity, long-term UV exposure, and histopathological and laboratory findings. Discoid LE was diagnosed based on clinical, histopathological, and direct immunofluorescence (DIF) findings. Care was taken to choose clinically well-demarcated, coin-shaped, squamous plaques with adherent scales, follicular plugging, intermittent telangiectasia and peripheral hyperpigmentation. In older lesions, those with central atrophy and hypopigmentation were chosen. Typical histopathological features of DLE included orthokeratotic hyperkeratosis, follicular plugging, epidermal atrophy, dyskeratosis, basal cell degeneration, basement-membrane thickening, and perivascular and periadnexal mononuclear cell infiltrate. During the DIF examination, granular deposition of IgG and C3 at the dermoepidermal junction was also investigated. Exclusion criteria were age younger than 18 years, absence of discoid lupus diagnosis, and lack of medical records.

## Statistical analysis

Data were analyzed using SPSS for Windows version 15.0 (SPSS Co., Chicago, IL, USA). Normal distribution of data was analyzed using histogram plots and the Kolmogorov-Smirnov test. Descriptive data were expressed as mean, standard deviation (SD), median, frequency, and percentage. Nonparametric groups were compared using Mann-Whitney *U* and Kruskal Wallis tests. Categorical variables were compared using the  $\chi^2$  test and Fisher's exact test. A *p*-value of < 0.05 was considered significant.

## Results

The study included 132 patients comprising 67 (50.8%) men and 65 (49.2%) women. Of the 132 patients, 72 (54.5%) were either unemployed or housewives. Mean age was 43.2 ±14.1 years and mean age at onset was 39.3 ±14.1 years (Table 1).

A family history was found in 3.8%, SLE was detected in 5.3%, and photosensitivity was revealed in 50.0% of the patients. ANA positivity was found in 23.7%, a history of smoking was revealed in 61.4%, and chronic sun exposure was detected in 42.4% of the patients. Hypertension (HT) was the most common comorbidity (18.2%), followed by diabetes mellitus (DM) (6.8%) and hyperthyroidism (2.3%). No comorbidity was detected in 72.7% of the patients (Table 2).

An analysis on the treatment methods indicated that 99.2% of the patients underwent treatment with sun-screen sun protection factor (SPF), 90.9% underwent

topical corticosteroid therapy, 56.8% underwent hydroxychloroquine therapy, 19.7% underwent topical calcineurin therapy, and the remaining 2.3% received other therapies. On the other hand, hairy skin involvement was detected in 22.7%, facial involvement was detected in 78.8%, and whole body involvement was detected in 15.2% of the patients. A positive DIF finding was detected in 86.4% of the patients (Table 3).

A comparison between gender and family history, site of involvement, and treatment methods revealed that ANA positivity was higher in women than in men (32.3% vs. 15.2%) ( $p = 0.021$ ). However, no significant difference was found between gender and other variables ( $p > 0.05$ ) (Table 4).

The 12-year incidence rate over the period 2005–2017 was 12.62 cases per 100,000 population and the estimated annual incidence rate was 1.05 cases per 100,000 population.

**Discussion**

The results indicated that DLE has a low incidence in the eastern part of Turkey and, contrary to common belief, there is no female preponderance in DLE. To our knowledge, there are very few studies in the literature reporting on the incidence rate of DLE. Durosaro *et al.* reported the annual incidence rate of DLE as 4.3/100,000 population while Gronhagen *et al.* found a rate of 1.05/100,000 population. In our study, we found a rate of 1.05/100,000 population, which was lower than that of other studies [8, 9].

**Table 3.** Treatment methods and sites of involvement

Parameter		Number	Percentage
SPF	No	1	0.8
	Yes	131	99.2
Topical corticosteroid	No	12	9.1
	Yes	120	90.9
Hydroxychloroquine	No	57	43.2
	Yes	75	56.8
Topical calcineurin inhibitor	No	106	80.3
	Yes	26	19.7
Others	No	129	97.7
	Yes	3	2.3
Hairy skin*	No	102	77.3
	Yes	30	22.7
Face*	No	28	21.2
	Yes	104	78.8
Whole body*	No	112	84.8
	Yes	20	15.2
DIF test	Negative	18	13.6
	Positive	114	86.4

SPF – sun protection factor, DIF – direct immunofluorescence, \*involvement.

Studies have indicated that DLE is more common in women than in men. Of note, the female-to-male ratio was reported as 3.2/1 by Koskenmies *et al.* and as 5/1 by both Jacyk and Damisah, and Insawang *et al.* In our

**Table 4.** Gender-based comparison of family history, site of involvement, and treatment methods

Parameter		Gender				P-value
		Male		Female		
		n	%	n	%	
Family history	No	65	97.0	62	95.4	0.678 <sup>b</sup>
	Yes	2	3.0	3	4.6	
Systemic lupus erythematosus	No	65	97.0	60	92.3	0.270 <sup>b</sup>
	Yes	2	3.0	5	7.7	
Photosensitivity	No	38	56.7	28	43.1	0.117 <sup>a</sup>
	Yes	29	43.3	37	56.9	
ANA positivity	No	56	84.8	44	67.7	<b>0.021<sup>a</sup></b>
	Yes	10	15.2	21	32.3	
Hairy skin*	No	50	74.6	52	80.0	0.461 <sup>a</sup>
	Yes	17	25.4	13	20.0	
Face*	No	17	25.4	11	16.9	0.235 <sup>a</sup>
	Yes	50	74.6	54	83.1	
Whole body*	No	55	82.1	57	87.7	0.369 <sup>a</sup>
	Yes	12	17.9	8	12.3	
Duration of disease		2.0	0.1–11.0	2.0	0.1–12.0	0.080 <sup>c</sup>

<sup>a</sup> $\chi^2$  test; <sup>b</sup>Fisher's exact test; \*involvement; for quantitative values, median was used instead of n and minimum-maximum values were used instead of %; <sup>c</sup>Mann-Whitney U test.

study, the incidence of DLE in men and women was almost equal, which, contrary to common belief, suggests that there is no female preponderance in DLE [6, 10, 11].

Discoid LE is mostly seen between the ages of 40 and 60 years and the mean age at onset was reported to be 42 years by Koskenmies *et al.* and 38 years by Ng *et al.* [10, 12]. Similarly, we also found that the mean age at onset was 39.3 years, which was consistent with the literature. Taken together, these findings indicate that DLE has a peak incidence in the third to fourth decades of life. Nevertheless, apart from these studies, there are also several pediatric studies with large patient series [13].

Discoid LE mostly involves the face and hairy areas of the skin. Tang *et al.* reported that the face was the most common site of involvement as opposed to hairy areas of the skin in the study by Fahad *et al.* [7, 14–16]. In our study, the rate of facial involvement was higher than that of other sites, which implies the presence of photodermatosis.

Literature reviews indicate that there are contradictory rates of photosensitivity in patients with DLE. Insawang *et al.* reported a rate of 14.6% and 71.3% in two different studies while Callen reported a rate of 87%. In contrast, we found a rate of 50%, which was an average rate when compared to the rates reported in the literature. All these rates imply that the incidence of photosensitivity is seriously high in DLE and thus protection from sunlight is highly important in patients with DLE [6, 10, 17].

Although DLE is known to be aggravated by sun exposure, chronic sun exposure is not mentioned in most studies. In our study, chronic sun exposure was detected in 42.4% of the patients, which suggests the importance of the cumulative effect of sunlight exposure in DLE. Moreover, studies have also shown that UVB could be a more important factor than UVA in DLE [7].

Some cases of DLE may progress to SLE or may coexist with both DLE and SLE. The rate of progression to SLE was reported as 20.7% by Insawang *et al.* and as 17% by Tang *et al.* On the other hand, the rate of coexistence of DLE and SLE was reported as 9.8% by Grönhagen *et al.* and 6.5% by Callen [6, 7, 14, 17]. In our study, we found that the rate of coexistence was 5.3%, which was lower than the rates reported in the literature. However, taken together, all these rates imply that only a low rate of patients with DLE progress to SLE. Moreover, compared to acute and subacute LE, DLE appears to provide protection against progression to SLE since the acute and subacute forms of DLE lead to high rates of progression [1, 6, 7].

Antinuclear antibodies have been shown to be highly important in patients with DLE and to be a potential indicator of progression to SLE. ANA positivity was detected in 16.1% of the patients evaluated by Fahad *et al.* and in 67% of the patients evaluated by Tang *et al.* [14, 15]. In our study, ANA positivity was detected in 23.7% of the patients. Although this rate was similar to the rate reported by Fahad *et al.*, it was lower than the rate reported

by Tang *et al.*, which probably resulted from the relatively smaller patient series evaluated by Tang *et al.* Moreover, the rate of progression to SLE (5.3%) and ANA positivity (23.7%) in our study suggests that the ANA positivity in DLE is not associated with the presence of SLE. However, the significantly high rate of ANA positivity in our female patients suggests that this parameter is likely to differ between genders.

There are contradictory rates of family history in patients with DLE in the literature. While Sampaio *et al.* found a rate of 11.8%, Insawang *et al.* reported that no family history was detected in any patient [6, 13]. In our study, however, a family history was detected in 5 (3.8%) patients, which suggests the presence of a genetic tendency.

Smoking has been implicated in the etiopathology of DLE in some studies. Miot *et al.*, for instance, reported that 84.2% of patients and 33.5% of control subjects had a history of smoking, also noting that the rate of smoking was higher in the patients with DLE and smoking is a leading factor aggravating DLE [18]. Similarly, Böckle *et al.* found that there was a high rate of smoking among patients with DLE and lupus erythematosus tumidus (LET) [19]. In line with the literature, the smoking rate in our patients was 61.4%, which suggests that complete cessation of smoking is imperative in patients with DLE.

The first step in the treatment of discoid lupus erythematosus is high potency topical corticosteroids. In a previous study, topical steroid treatment was given to all patients with discoid lupus. However, some of the patients did not respond to treatment [6]. In another study, 60% of patients received topical corticosteroid treatment [20]. In our study, 90.1% of patients received topical corticosteroid therapy. This was consistent with the literature.

Our study was limited since it was a single-center retrospective study. Additionally, no anti-double stranded DNA (anti-dsDNA) test was performed in some patients due to technical difficulties, and the vitamin D level was not measured in any patient.

## Conclusions

Discoid LE, though identified many years ago, remains unelucidated. Moreover, there are very few studies in the literature reporting on DLE. The results of this study indicated that the estimated annual incidence rate of DLE in the eastern part of Turkey is 1.05 cases per 100,000 population and smoking and chronic UV exposure are important risk factors for DLE. Moreover, although ANA positivity was high in our patients, the rate of progression to SLE was remarkably low. The results also showed that, contrary to common belief, there is no female preponderance in DLE. Further prospective multicenter studies with large patient series are needed to better understand the clinical features of DLE.

**Conflict of interest**

The authors declare no conflict of interest.

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